



Olfactory Schwannomas – A Case Series with Long-Term Outcomes

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Abstract

Background: Olfactory schwannomas are rare tumors arising from olfactory groove whose pathogenesis is still not understood. Only a few case reports are available, and their long-term outcome remains vague. We report the surgical outcome of five patients of olfactory schwannomas with one having over twenty years of follow up.

Methods: A retrospective review of all intracranial schwannomas operated at our centre over last 20 years was done from which five primary cases of tumor located in the subfrontal/anterior cranial fossa floor were analysed. Also, a systematic review of pertinent previous English language literature was conducted using PubMed.

Results: Four of five olfactory schwannoma cases were seen in young females. Anosmia/hyposmia and symptoms of raised intracranial pressure were the most common presentations. Radiologically, these tumors were solid cystic arising from olfactory groove having areas of haemorrhage and necrosis. Thet of the lesions (5 out of 6 surgeries: one recurrent) were operated via the sub-frontal approaches, either lateral or anterior. We noted that the presence of bony erosion warrants robust anterior cranial fossa floor repair, as postoperative cerebrospinal fluid rhinorrhoea was present in two out of three patients. These tumors had good plans of cleavage with the surrounding brain parenchyma but have the surgical challenge of identification and prevention of injury to anterior cerebral arteries. Aggressive resection could be achieved in all patients and long-term follow-up was favourable. Also, when respected completely, they do not require adjuvant therapy.

Conclusion: Though other anterior cranial fossa tumors are close differentials, olfactory schwannomas are unique in terms of clinical behaviour, histopathological profile and surgical excision philosophy, the latter being warranted by more frequent repair of the skull base. More evidence is required to support appropriate theories of origin of these tumors.

Keywords: Olfactory Schwannomas; Subfrontal Schwannoma; Anterior Cranial Fossa Tumors; Schwannomas

Abbreviations

CSF: Cerebrospinal Fluid; ACA: Anterior Cerebral Arteries;
ICP: Intracranial Pressure; ACF: Anterior Cranial Fossa.

Introduction

Olfactory schwannomas are often described as olfactory groove schwannomas, subfrontal schwannomas or anterior cranial fossa schwannomas and represent a rare form of skull

base tumors of whose pathogenesis is still debatable. These are enigmatic entities in neurosurgery as olfactory nerve does not have a Schwann cell layer and no other structure in anterior cranial fossa –floor have them. The rarity of this tumor, coupled with clinico-radiological features like those of other regional tumors, often results in misdiagnosis prior to surgical pathologic examination. Misdiagnosis may lead to an inappropriate surgical approach and excision. To date, very few cases have been reported. This article fulfills the need for us to know the long-term outcome following surgery. This article enumerates the post-operative fate of subfrontal schwannomas, underscoring a gratifying long-term response to surgical intervention and their benign nature.

Material and Methods

Following Institutional Ethics committee approval, we performed a retrospective review of all histologically proven intracranial schwannoma managed surgically at our institution between 1999 and 2019. Among these cases, individuals were identified from a patient database system using operative notes to search for those tumors located in the

subfrontal/anterior cranial fossa (ACF) floor. The patient's charts and operative records, radiological images from PACS and outpatient department follow-up sheets were analysed to collect data pertaining to patient demographics, unique clinical features, radiological findings, operative approaches, findings and post-operative management, histopathological examinations and outcome/prognosis of these rare tumors. Gross-total resection was defined as the absence of residual tumor on postoperative contrast imaging. A review of the previous literature from PubMed using keywords like subfrontal schwannoma, olfactory schwannoma, anterior cranial fossa schwannoma was done focusing on the origin of these schwannomas and its proposed etiopathogenesis.

Results

Of 1236 consecutive cases of intracranial schwannomas surgically treated in our institution over last 20 years, only six cases (five primary and one recurrent case) were found to be in the subfrontal/ACF floor accounting for less than 0.5% of intracranial schwannomas (Table 1).

c	Age/Sex	Presentation	Anosmia/Hyposmia	Size (mm)	Craniotomy	Approach	Origin of Tumor	Bony Erosion	Complications	Follow Up
1	25/F	Headache	Yes	50x50	Left frontal	Transcortical	Olfactory Groove	Absent	Nil	20 Years
2	45/M	Right Eye Proptosis	Yes	73x64x89	Bifrontal	Anterior Subfrontal	Olfactory Groove	Present	CSF rhinorrhoea	16 Years
3	36/F	Seizures	Yes	57x51x31	Bifrontal	Anterior Subfrontal	Olfactory Groove	Present	Nil	12 Years
4	39/F	Headache (Recurrence)	Yes	51x50x69	Bifrontal	Anterior Subfrontal	Olfactory Groove	Absent	Nil	8 Years
5	49/F	Headache	Yes	76x60x86	Pterional	Lateral Subfrontal	Olfactory Groove	Present	CSF rhinorrhoea	2 Years
6	24/F	Headache	Yes	55x49x43	Pterional	Lateral Subfrontal	Olfactory Groove	Absent	Nil	6 Months

Table 1: Patient characteristics.

We found that these tumors tend to occur in young females of age group 24 to 49 years. All the 5 primary cases operated for subfrontal schwannomas, had anosmia/hyposmia while headache and raised intracranial pressure (ICP) symptoms were present in 4/5 patients. One patient presented with seizure and another one presented with proptosis along with features of raised ICP. Radiologically, these tumors were well defined as predominantly solid-cystic arising from midline anterior cranial fossa floor with heterogenous contrast enhancement Figure 1. These lesions were hypo to innocents on T1WI and hetero-hyperintense on T2WI sequences with areas of necrosis and hemorrhage. These tumors were

associated with significant perilesional edema in five of six cases and presented when of significantly large size. Bony erosions into paranasal sinuses and orbit were seen in fifty percent of patients. Advance sequences were available for only 2 patients which revealed blooming in susceptibility weighted images suggestive of intra lesional hemorrhages, as corresponding CT images did not show any calcifications. There was no evidence of any diffusion restriction in both the patients. Angiograms were performed in one patient that revealed tumor blush in anterior cranial fossa with arterial feeders from poster-superiorly displaced anterior cerebral arteries (ACA). There was no narrowing of the ACA noted.

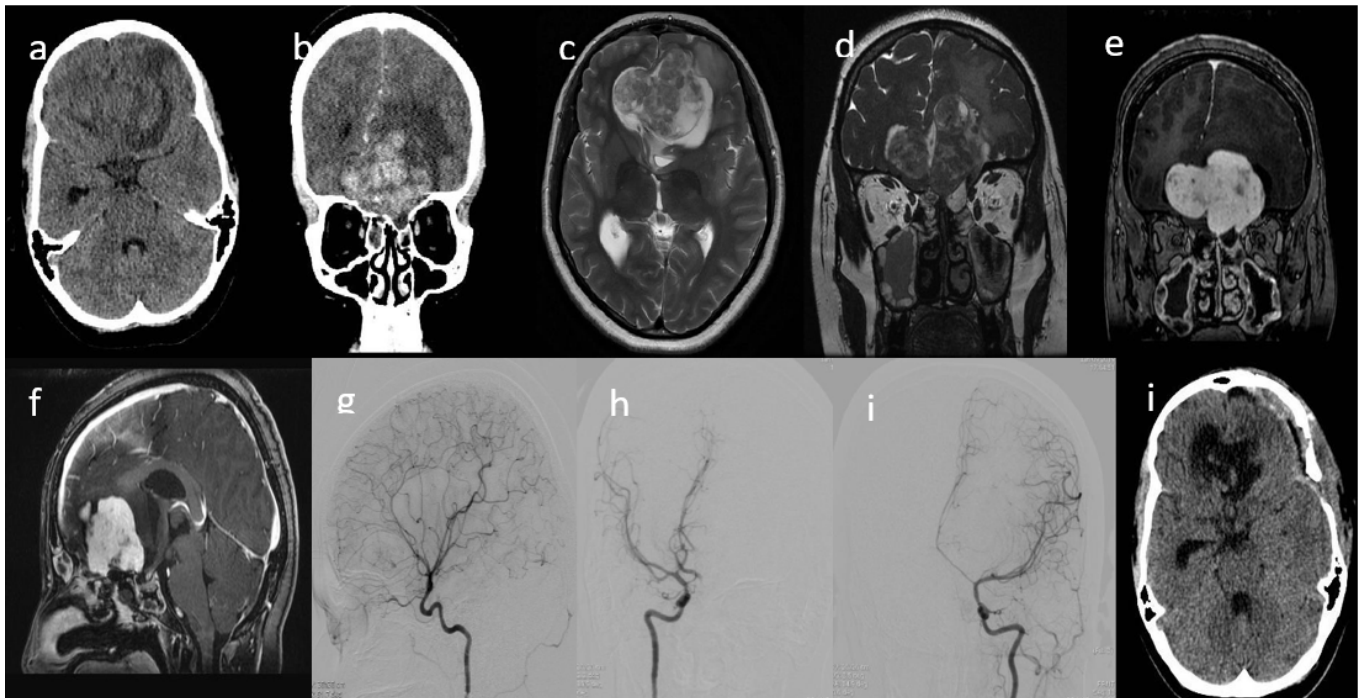


Figure 1: Axial plain (a) and coronal contrast (b) CT cuts show iso to hypodense extraaxial lesion arising from anterior cranial fossa with heterogeneous contrast enhancement and mass effect and edema in adjacent parenchyma. T2WI axial (c) and coronal (d) cuts show predominantly solid, iso to hyperintense lesion with interspersed cystic areas and foci of micro-hemorrhages. On coronal (e) and sagittal (f) contrast images, a well-defined lobulated heterogeneous intensely enhancing mass in anterior cranial fossa with no dural tail is seen. The lesion appears to arise from left cribriform plate. Angiograms (g, h and i) show mild tumor blush in anterior cranial fossa with arterial feeders from ACA along with superior displacement of orbitofrontal branch of right ACA and postero-superior displacement of the right A2 ACA. Left A1 ACA is stretched and displaced superiorly and laterally while left A2 ACA is displaced to right side. Axial CT (j) shows postop changes with complete resection of the tumor.

Of 6 patients, the first patient was operated on by frontal craniotomy and transcortical approach probably because of predominantly cystic nature of this tumor, latter 2 cases and the recurrent tumor was operated by bifrontal craniotomy and anterior subfrontal approach. The last 2 cases were operated by pterional craniotomy and lateral subfrontal approach. Gross total decompression was achieved with all the approaches; however, recurrence was noted in patient operated by transcortical route. At surgery, these tumors were predominantly well demarcated, yellowish-greyish solid cystic, vascular, amenable to suction and had good planes of cleavage from parenchyma and basal dura Figure 1.

These tumors were adhered to olfactory groove/cribriform plate region with olfactory nerve/tract could not be separately identified from the tumor and could not be preserved. ACAs were pushed posteriorly and there was no vessel injury or encasement. Bony erosion was evident in 3 cases of which 2 had cerebrospinal fluid (CSF) rhinorrhea in post-operative period that required surgical exploration and repair with lumbar drain.

Histopathologically, these schwannomas were characterised by the presence of spindle cells disposed of hypercellular Antoni A and hypocellular Antoni B areas Figure 1. In the Antoni A areas, cells were arranged in compact fascicles and Schwannian whorls with or without nuclear palisades (Verocay bodies). In the Antoni B areas, the cells were dispersed in the loose microcystic stroma.

Cystic change, lymphocytic and foamy macrophage infiltration, hyalinised blood vessels, haemosiderin pigment, and degenerative nuclear atypia were common. These tumors were diffusely positive for S-100 protein. In our series, all the cases showed the biphasic pattern with a predominance of Antoni A morphology with Verocay bodies being present in 4 cases.

The follow-up ranged from 6 months to 20 years. There were no new deficits or new infarcts (venous/arterial) in any of the patients in postoperative period. All patients were independent for activities of daily living and have returned to their normal life. Detailed neuropsychological

evaluation was not performed in any patients however, patients reported improvement in their memory, ability to concentrate and social interaction. As per institute protocol, antiepileptic drugs were gradually tapered and stopped in all patients after 2 years of seizure-free interval. During the period of follow-up, only one patient developed recurrence after 13 years, who was managed with surgical exploration. None of the patients required any form of adjuvant therapy.

Discussion

Schwannomas are usually slow-growing benign neoplasms that originate from Schwann cells of peripheral or cranial nerves. Intracranial schwannomas represent about 6 to 8% of all intracranial tumors and arise most commonly from the vestibular branch of the eighth cranial nerve or less commonly from the fifth nerve, seventh nerve, and lower cranial nerves. The olfactory and optic nerve lack a Schwann cell layer and thus are not prone to tumors from these nerves. However, mysterious olfactory schwannomas are reported and to date, only 67 cases have been described in literature [1,2]. We add a series of 5 such cases that fulfill the need for information on longterm outcome following surgery for these subfrontal/olfactory schwannomas. We also describe the only recurrence reported.

The origin of these kinds of lesions is still ambiguous. Several hypotheses based on developmental and non-developmental theories have been proposed to explain their genesis [3].

The developmental hypotheses suggest these tumors may derive from the mesenchymal pial cells that transform into ectodermal Schwann cells, or they may arise from multipotent mesenchymal cells.

Another theory suggests that neural crest cells migrate within the substance of the central nervous system forming foci of Schwann cells (schwannosis) [1,4].

The non-developmental theories postulate that subfrontal schwannomas may arise from the Schwann cells of the adjacent structures, such as the perivascular nerve plexus, the meningeal branches of the trigeminal and anterior ethmoidal nerves or the fila olfactoria, a Schwann-cell sheath that extends 0.5 mm beyond the olfactory bulb [3,5].

Adachi et al have classified subfrontal schwannomas into two main types: olfactory site schwannomas and "other than olfactory site" schwannomas, arising from nonolfactory sites. In our series, all the schwannomas originated from the olfactory groove or cribriform plate [6].

Okamoto et al. reported originating of schwannoma between the outer endosteal layer and the inner meningeal layer of

the dura mater. They proposed that these schwannomas could arise from the Schwann cells of nerves that penetrate through the dura mater or from Schwann cells associated with fila olfactoria [7]. Quick et al also supported similar mechanism of origin. Although, our series shows these tumors were adherent to the cribriform plate/olfactory groove and olfactory nerves/tracts could not be seen separately, we could not find enough evidence to support either of the theories, probably because, all our tumors were large in size distorting the normal structures [8]. Similarly, presence of anosmia/hyposmia in almost half of the reported cases, and the predominant midline occurrence, favours origin of these schwannomas from the region of olfactory groove but could not prove either of the theories [9,10].

These schwannomas tend to occur in younger patients than those affected by schwannomas in more common sites with female predominance as seen in our series. This was contrary to most of the previous literature which were case reports and could now tilt the balance towards female predominance.

Various radiological mimickers of extra-axial masses arising in the anterior cranial fossa and midline with extension to surrounding structures include meningioma, esthesioneuroblastoma, ethmoid carcinoma, lymphoma, and metastatic disease. However, the T2*WI MRI sequence, showing multiple foci of low signal intensities (due to micro bleeds) and the presence of bone scalloping on CT, with absence of a dural tail sign, hyperostosis and more aggressive bony destruction with an invasion of the paranasal sinuses point towards schwannoma. Extension into sinuses/ orbit, which is commonly reported, was also seen in our series [11,12].

Surgery is the main therapeutic modality, and complete excision entails good prognosis and low recurrence rates compared with other ACF tumors like meningiomas and extensive fungal granulomas. Surgical strategy includes gross total excision while preserving maximum function. The approach can be either lateral subfrontal through pterional/ frontal craniotomy or anterior subfrontal via bifrontal craniotomy depending upon the location and extension of the tumor and surgeon's preference. The former can be preferred as these tumors have good planes with surrounding parenchyma and dura, amenable to suction, thereby avoiding bilateral frontal retraction injuries. Our experience with these schwannomas has put light on the surgical challenges in tackling these large

tumors that include abnormal position/displacement of major vessels like ACAs and bony erosion or extension to orbit and paranasal sinuses rising the incidence of CSF rhinorrhoea. Therefore, the surgical management should also include robust repair mechanism of anterior cranial

fossa floor following surgical excision. Hence, a preoperative suspicion of olfactory schwannomas can help deciding appropriate surgical strategy and improve the outcomes.

Our study of histologically proven olfactory schwannomas gives long term follow up results that were aggressively treated. Outcomes have been favourable with excellent prognosis and without any need for adjuvant therapy.

Conclusion

Olfactory schwannomas are very rare tumors and the debate about the origin of olfactory schwannomas is still open. Aggressive surgical excision with ACF base repair is the main modality of treatment. Olfactory schwannomas should be considered as differential in the case of anterior cranial fossa floor tumors as they have a favourable prognosis and good long-term outcomes. They do not require any form of adjuvant therapy when respected completely.

Compliance with Ethical Standards

Funding

No funding was received for this research.

Conflict of Interest

The authors declare that they have no conflict of interest.

Ethical Approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional ethics committee.

Informed Consent

Individual patient consent was not required as the study was retrospective one and was undertaken with local institutional approval.

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